Soft tissue myoepithelial cell carcinoma

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Dear Sir,

Myoepithelial cell carcinoma, first described by Stromeyer et al in 1975, is defined as a malignant carcinoma that primarily arises from the parotid gland;[1] it may also arise from the mammary glands. In 1991, myoepithelial cell carcinoma was added to the second edition of the World Health Organization's classification of malignant salivary gland tumours.[2] Soft tissue myoepithelial cell carcinoma (STMC) is rare compared to its counterparts arising from the salivary glands.

We herein describe the case of a 39-year-old man who was referred to Singapore General Hospital, Singapore, for swelling of the left thigh that was initially diagnosed as extraosseous chondrosarcoma. However, repeat biopsy results indicated the diagnosis of STMC. The patient had undergone two cycles of neoadjuvant chemotherapy. Subsequent repeat magnetic resonance imaging showed that the tumour, which arose from the vastus medialis, had increased in size from approximately 20.0 cm × 18.8 cm × 27.3 cm to about 23.2 cm × 20.1 cm × 32.9 cm. Surgical resection options were discussed, but the patient declined surgery.

STMC is rarely reported compared to carcinoma of the parotid gland. Patients who were diagnosed with STMC ranged from 3–83 years of age, with a mean age of 40 years and a slight male predilection.[3] Nearly two-thirds of reported cases were for tumours arising from the extremities (38% lower and 27% upper extremity); the remainder involved the head and the neck regions (16%), trunk (13%), and visceral soft tissue (6%).[4] Approximately 60% of these tumours are subcutaneous in origin and 40% occur in deep soft tissue (i.e. intramuscular or subfascial).[4]

In our patient, we experienced difficulty in interpreting the histological diagnosis. This is not uncommon as, due to histological polymorphism, STMC poses different challenges. Several entities share similar characteristics with STMC. Hence, immunohistochemistry investigation plays a vital role in differentiating this type of tumour. The neoplastic myoepithelial cells express the epithelial markers AE1/AE3 (90%) and EMA (60%), and myoepithelial markers S-100 protein (89%), calponin (87%), glial fibrillary acidic protein (46%) and smooth muscle actin (36%).[5]

Regarding the genetic study of myoepithelial cell carcinoma, the study by Antonescu et al, which included 66 cases, showed that the EWS RNA-Binding Protein 1 (*EWSR1*) gene rearrangement was a common event in myoepithelial tumours arising outside of salivary glands, as found in 45% of cases.[6] The *EWSR1* gene is located on chromosome band 22q12 and encodes a promoter-specific transactivator.[7] *EWSR1* gene rearrangement is also associated with several neoplasms such as Ewing's sarcoma, clear cell sarcoma, myxoid liposarcoma and extraskeletal myxoid chondrosarcoma.[6] However, a study by Rekhi et al[7] reported that only 50% of STMC cases expressed *EWSR1* gene rearrangement; similarly, our patient did not exhibit *EWSR1* gene rearrangement. This indicated that *EWSR1* gene rearrangement may not necessarily present in general cases of STMC. Another study by Aparicio et al showed that a patient diagnosed with *EWSR1*-negative subsequently passed away due to multiple metastasis,[8] suggesting a poor prognosis and the need for close follow-up.

Complete surgical resection is recommended for treatment of myoepithelial cell carcinoma. As our patient declined surgery, he only received chemotherapy, which failed to control the tumour locally. A recent study has reported positive clinical outcomes through the use of a treatment strategy involving a combination of aggressive local control, chemotherapy and radiotherapy.[9] However, as the study mainly involved paediatric patients and was limited by its small size, further long-term patient follow-up is required. Two studies reported a 42% risk of local recurrence of the tumour and that a further 32%–52% had metastasised.[3,10] This indicates that myoepithelial cell carcinoma can have an aggressive clinical course. Common reported sites of metastasis include the lungs, bone, lymph nodes and soft tissue.[10] Even benign myoepithelioma tumours have a reported 20% risk of local recurrence.[3]

In conclusion, due to the rare nature of STMC, it remains a diagnostic challenge. However, the implementation of immunochemistry and genetic typing as diagnostic tools will help to accurately differentiate STMC from other tumours and, in turn, improve prognosis. As there is currently no general consensus on the management of STMC, a combination strategy may be the key to the treatment of this particular tumour.

Yours sincerely,

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